SHORT REPORT

Rub epilepsy: a somatosensory evoked reflex epilepsy induced by prolonged cutaneous stimulation

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Abstract

To delineate rub epilepsy—a type of reflex epilepsy induced by prolonged or repetitive cutaneous stimulation in a circumscribed area of the body—three cases are presented, as well as one of tooth brushing epilepsy for comparison. In all three cases of rub epilepsy, cutaneous stimuli in a particular body area on the left side initially induced a sensory jacksonian march in the middle of, or in close vicinity to, the trigger zone, which led to subsequent unilateral tonic contractions with intact consciousness. By contrast, a motor jacksonian seizure without sensory aura was induced in the patient with tooth brushing epilepsy. A review of cases with rub epilepsy, including those in this paper, disclosed a striking consistency in clinical manifestations. The symptomatology of the induced seizures indicates a propagation of epileptic discharges from the postcentral gyrus to the supplementary motor area. Rub epilepsy is proposed as a separate clinical entity, clearly demarcated from other somatosensory evoked reflex epilepsies such as startle and tooth brushing epilepsy.

(*J Neurol Neurosurg Psychiatry* 2001;**70**:541–543)

Keywords: somatosensory evoked reflex epilepsy, startle epilepsy, tooth brushing epilepsy

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Received 15 May 2000 and in revised form 29 November 2000
Accepted 19 December 2000

As long ago as 1863, John Hughlings Jackson described a case in which touching the thumb would bring on a fit.[1] Later, in 1910, Woodcock and Edin described a boy who went into epileptic fits repeatedly when his sister removed the stocking from his right leg.[2] However, until recently, somatosensory evoked epilepsy without an element of surprise has been confused with other types of somatosensory evoked epilepsy such as startle epilepsy. We have named the first type of somatosensory evoked epilepsy as "rub epilepsy", because a long or regular series of tactile contacts are usually the most effective means of eliciting a seizure. Herein, three cases of rub epilepsy, treated at Kansai Regional Epilepsy Center, are presented, along with a case of tooth brushing epilepsy as a comparison. Written consent was obtained from each patient before the report.

Case reports

CASE 1

A 35 year old woman began to have seizures at 14 years of age. The first attack came suddenly with a sensation of pressure in the left shoulder, immediately followed by secondary generalisation. Since that incident, paroxysmal dysaesthesia has occurred daily in that area. At the age of 17, the patient noticed that she could trigger seizures by repeatedly patting her left shoulder. After the stimulation, she would feel pressure in this region that would then extend down the left arm to the tip of her fingers. This was followed by either a tonic contraction of the left arm with a simultaneous leftward movement of the head, or by prolonged localised jerks of the fingers of the left hand. At the age of 35, she was transferred to a university hospital, because a generalised seizure had recurred after a 21 year interval. Occasionally, paroxysms of sudden atonia of the left limbs were also noted. She was diagnosed as having psychogenic seizures, because of the bizarre symptoms. After a reduction of antiepileptic drugs, spontaneous tonic contractions of the left limbs occurred every 15 minutes. Due to this cluster of seizures, the patient was referred to us. A neurological examination was normal, with no intellectual deficit found, and MRI showed no abnormality. After raising the level of phenytoin, the tonic seizures disappeared. An interictal EEG showed no epileptiform discharge, whereas an ictal EEG showed low amplitude fast rhythm activities over the central and parietal region on repeated tapping of the left shoulder, followed by widespread moderate amplitude fast rhythm activities when the tonic motor signs appeared.

CASE 2

A 21 year old woman first had a paroxysmal tickling sensation at the age of 12 years. The sensation was initially localised in the left knee, and then spread to the entire left half of her body. This was occasionally followed by a tonic contraction of the left limbs. Afterwards, she incidentally discovered that she could induce seizures by rubbing her left leg. As an interictal EEG demonstrated no epileptiform discharge and because this peculiar complaint was unfamiliar to the neurologist who treated her, she was suspected as having psychogenic seizures

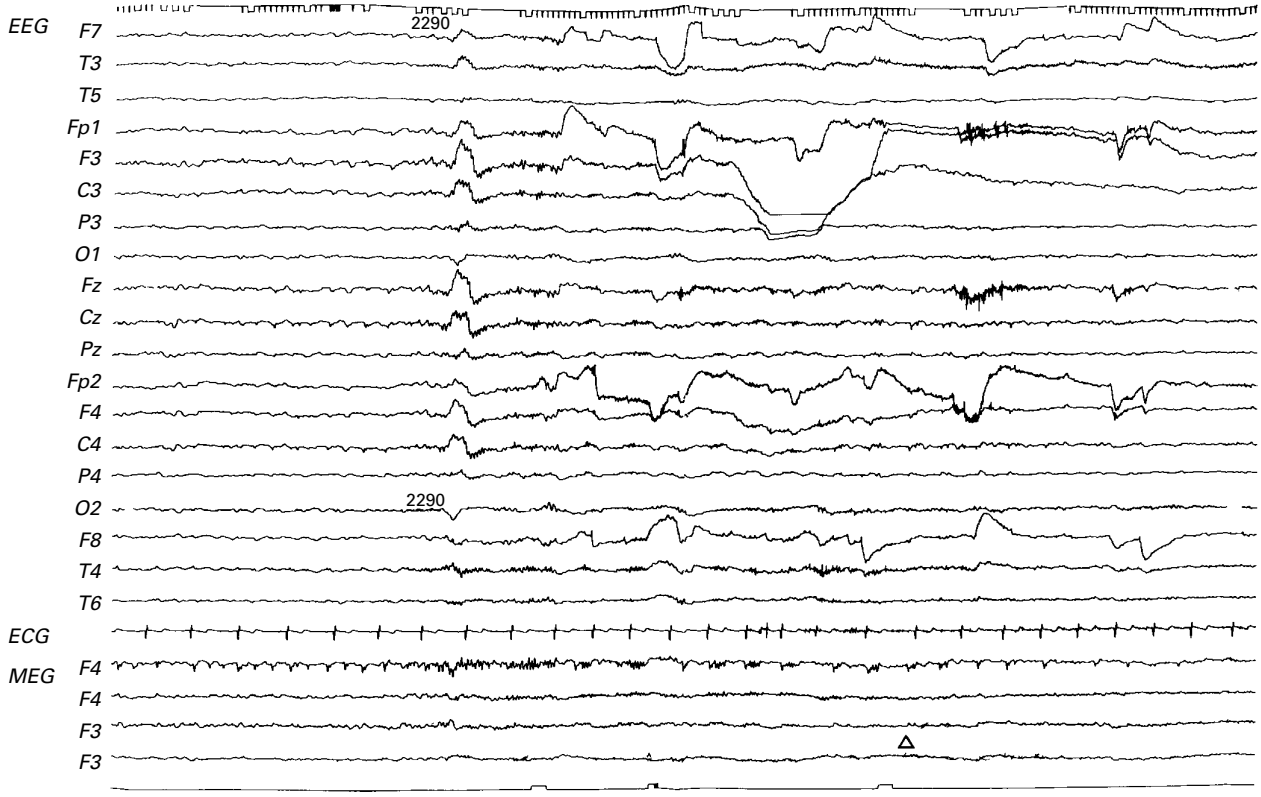

Figure 1 Appearance of a tickling sensation localised in the left leg. Whereas an ictal EEG showed only diffuse fast rhythm activity, simultaneous MEG recording demonstrated spike trains in the right frontal region, which preceded the ictal EEG discharge.

and referred to us. The neurological examination was normal, with no intellectual deficit found, and MRI showed no abnormality. The interictal EEG was also normal, whereas the ictal EEG showed diffuse low amplitude fast rhythm activities. An ictal magnetic encephalography (MEG) recorded at Shizuoka Higashi National Epilepsy Center demonstrated repetitive spikes, followed by polyspikes localised in the right frontal region (figure 1).

CASE 3

A 44 year old patient first began to experience seizures at 18 years of age. They began with an impression of scratchiness in the left thorax, which then spread to the left vertex region and changed into a throbbing sensation. At the age of 28, motor manifestations with tonic contractions of the left arm and turning of the head to the right began to follow the somatosensory attacks. At the age of 31, the patient noted, incidentally while receiving a haircut, that the throbbing sensation in the left vertex could be provoked by the rubbing of a well circumscribed area in the head, which was later discovered to correspond to the location of the C3 electrode. He found that he needed to rub this area for more than 10 seconds to provoke the sensation, which was often followed by the habitual tonic contractions of the arm and head turning. Seizures occurred every day, but the patient noticed that after a self induced seizure, the trigger zone was no longer effective in producing further seizures for several hours, which permitted him to continue his activities and to tolerate hair cutting. A sudden startle did not trigger seizures. At 34 years of age, his seizures were diagnosed to be psychogenic and antiepileptic drugs were discontinued. After that, they occurred spontaneously, without any provoking stimuli, every 10 to 15 minutes and became generalised, which led him to be transferred to our institute. A neurological examination was normal, with no intellectual deficit found. Brain MRI and interictal EEG examinations were also normal. After raising the blood concentration of phenytoin, even optimal stimuli could only rarely trigger seizures.

CASE 4

A 45 year old woman first began to have seizures at 3 years of age. They started with a twitching of the left corner of her mouth, and occasionally spread to the left neck or lower face, leading to a repeated turning of the head to the left and facial grimacing. By the age of 30, she was experiencing attacks every day, most of which occurred during sleep and would last for 30 to 60 seconds. Daytime attacks were provoked by brushing teeth or eating solid foods, and could be induced by brushing the upper or lower teeth on either side of her mouth. An EEG investigation demonstrated that trains of 2 per second spikes would appear after about 2 minutes of tooth brushing in the right central region.

Discussion

A review of amply documented cases of rub epilepsy,[3–5] including ours, disclosed a striking consistency in seizure symptomatology, when the mode of triggering was strictly limited to a prolonged rubbing of a well circumscribed area of the body. The induced seizures consisted of

*Table 1 Rub epilepsy cases**

Ref (1st author)		*Year of onset*	*Sex*	*Trigger zone*	*Induced seizures*†	*Frequency*
Goldie[4]		7	M	R face	R unilateral tonic	Daily
Foster[3]	Case 3	10	M	L palm	L unilateral tonic	Weekly
Vignal[5]	Case 1	7	M	L anterior thorax	L unilateral tonic	Daily
	Case 2	15	F	L iliac fossa	L unilateral tonic	Daily
Present study	Case 1	18	M	L anterior head	L unilateral tonic	Daily
	Case 2	14	F	L anterior shoulder	L unilateral tonic	Daily
	Case 3	12	F	L leg	L unilateral tonic	Daily

*The case reported by Rae[6] was excluded because only a painful stimulus to the hyperaesthetic area elicited seizures and emotional excitement was also reported to trigger them in that case. The case reported by Strauss[7] was also excluded, because rubbing the skin at any point on the right half of the body or a similar rubbing of symmetric points on both sides of the body simultaneously provoked seizures. †In all cases, sensory jacksonian seizures preceded motor seizures.

the following sequence. Initially, paraesthesia in the middle or in close vicinity to the trigger zone emerged, which spread to other body regions as a sensory jacksonian march, and finally unilateral tonic contractions ensued. This sequence was repeated regularly throughout the cases. Further, a refractory period was occasionally noted after self induced seizures, during which ensuing stimuli failed to trigger seizures for a certain period of time. This interesting feature of rub epilepsy is possibly due to a transient postictal surplus hyperpolarisation in response to preceding epileptic depolarisation. In table 1, we have summarised previous cases of typical rub epilepsy. Startle epilepsy and rub epilepsy have been confused for a long time. Indeed, both are triggered by tactile contact; however, the clinical constellations are different.[8 9] The first almost always occurs in patients with mental retardation, or in those having severe brain damage in the perinatal or early infantile period with resulting hemiplegia. Otherwise, diseases leading to generalised maldevelopment of the brain, such as Down's syndrome, cause startle epilepsy.[10] The induced seizures are almost always generalised tonic, although the tonic phase is at times so brief that only a sudden falling down can be noted. By contrast, rub epilepsy typically occurs in patients with neither mental nor neurological deficits. The relation between tooth brushing and rub epilepsy is more difficult to distinguish.[5] Although the gums certainly qualify as a well circumscribed body region, a scrutiny of the reported cases of tooth brushing epilepsy,[3 11–14] including ours, suggested that this type of epilepsy presents a different clinical constellation from that of rub epilepsy. Firstly, except for a case presented by Penfield, intraoral tactile stimulations which provoke seizures are not restricted to one side, but are bilaterally effective. In the nuclear group of rub epilepsy, the trigger zone exists only on one side of the body. Secondly, none of the patients with tooth brushing epilepsy mimicked the typical sequence of induced seizures in rub epilepsy—that is, initial sensory jacksonian seizures followed by unilateral tonic contraction. In 10 documented cases of tooth brushing epilepsy, including ours, motor jacksonian seizures were induced in eight patients[12 14] and complex partial seizures in the other two.[3 11] Thus, whereas seizure symptomatology of rub epilepsy indicates a propagation of epileptic discharges from the postcentral gyrus to the supplementary motor area,[15] the primary motor cortex is indicated as the area responsible for the symptoms in most cases of tooth brushing epilepsy. As motor symptoms originating from the supplementary motor cortex typically show only suppression during ictal EEG recordings,[16] results from scalp EEG recording in rub epilepsy have often been erroneously judged as normal and used as evidence to suggest a psychogenic nature. Further, patients with rub epilepsy are subject to constant psychological pressure, making them prone to the development of neurotic characteristics, because tactile stimulation to the trigger zone is only avoidable with the greatest possible care. Together with the rarity of rub epilepsy, these clinicoelectrical features can easily lead to misdiagnosis.

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